INTRAOCULAR JUVENILE XANTHOGRANULOMA (NEVOXANTHOGRANULOMA): A SURVEY OF 20 CASES*

BY *T. E. Sanders*, M.D.**

INTRAOCULAR JUVENILE XANTHOGRANULOMA is a rare skin disease of infants and young children which only recently has been recognized as the cause of an increasing number of unique iris lesions.

Although there are a few scattered reports of similar cases,[1-3] the skin lesion was first accurately described in 1909 by McDonaugh.[4] Three years later he added four more cases,[5] suggesting the term nevoxanthoendothelioma as descriptive of the lesion. He felt that they were a congenital endothelioma with a secondary fatty change. Not until 1936 did Senear and Caro[6] properly recognize the primary xanthomatous nature of the lesion. Helwig and Hackney[7] in 1954 reported 53 cases, emphasizing that a more desirable term for the lesion would be juvenile xanthogranuloma. There are a number of other reports on the skin lesion in the literature, probably totalling over 200 cases.[8-13]

In 1949, Blank, Eglick, and Beerman[14] reported in the pediatric literature the first case of intraocular involvement. The first such case in the ophthalmic literature was reported by Maumenee in 1956.[15] Since that time five additional cases with intraocular involvement have been added.[16, 17, 18]

The author has had the unusual opportunity to study three cases of this condition. On a review of the files of the Ophthalmic Pathology Registry of the Armed Forces Institute of Pathology 17 additional cases were found. These include four of the previously reported cases —the original case of Blank, Eglick, and Beerman,[14] that of Maumenee,[15] Newell's second case,[16] and Maumenee and Longfellow's second

*The original title of this paper, "Intraocular Nevoxanthoendothelioma (Juvenile Xanthogranuloma)" has been changed to conform to the more preferred term, as suggested by Dr. Hogan in his discussion.

**From the Department of Ophthalmology and the Oscar Johnson Institute, Washington University Medical School, St. Louis, Missouri.

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case.[18] Inasmuch as there is no series of intraocular involvement of this condition previously reported, it was thought a comparative study of these 20 cases was worthwhile (Table 1).

TABLE 1.

*Case**	*Sex*	*Age*	*Skin lesion*	*Hyphema*	*Glaucoma*	*Results*
1	M	2 mos.	Yes	Three	Yes	Enucleation—glaucoma
2	F	3 mos.	Yes	Yes	Yes	Enucleation—glaucoma
3	M	4 wks.	Yes	Yes	Yes	Enucleation—tumor
4	M	3 yrs.	Yes	Yes	Yes	Enucleation—tumor
5	F	9 mos.	?	?	?	Enucleation—tumor
6	M	6 wks.	?	Yes	Yes	Enucleation—tumor
7	F	2½ mos.	?	Yes	No	Enucleation—tumor
8	M	4 mos.	Yes	No	Yes	Enucleation—tumor
9	M	4 mos.	?	Three	Yes	Enucleation—tumor
10	M	4 mos.	Yes	Yes	Yes	Enucleation—tumor
11	F	7 mos.	Yes	Yes	Yes	Enucleation—tumor
12	M	7 mos.	?	Yes	Yes	Enucleation—tumor
13	F	11 mos.	Yes	Three	Yes	Skin biopsy—X ray
14	F	8 mos.	?	Yes	No	Enucleation—tumor
15	M	1 yr.	No	Yes	Yes	Iris biopsy
16	F	35 yrs.	?	No	No	Iris biopsy
17	F	6 mos.	No	Two	Yes	Iris biopsy
18	F	22 mos.	?	Yes	Yes	Iris biopsy
19	M	7 mos.	No	Yes	?	Enucleation—tumor
20	F	5 mos.	No	Yes	Yes	Enucleation—tumor

*With the exception of the author's cases (1, 2, and 3), the cases are listed in the order of accession in the Registry of Ophthalmic Pathology.

MATERIAL

The first recognized case (Case 8) in the files of the Registry of Ophthalmic Pathology is the original case of Blank, Eglick, and Beerman.[14] This was presented to the Ophthalmic Pathology Club in 1948 by Fry who had described the eye lesion in the original report. The next accessioned case was the author's first patient (Case 1), reported to the Ophthalmic Pathology Club in 1954. This case was presented with Slaughter and Klingberg as a clinicopathologic report at the American Academy of Ophthalmology in 1954 but not published. The next year three cases were reported at the Ophthalmic Pathology Club; by Maumenee (Case 10),[15] by Heath (Case 9) and the second case of Newell (Case 7),[16] which was presented to the Club by Theobald. It was later found that this case was sent to the Registry by Bothman in 1938 and was diagnosed at that time as malignant

melanoma of the iris. Since 1955 nine additional cases have been added by various contributors (Cases 12–20).

Recently, Drs. Rones and Zimmerman, in reviewing the iris tumor material in the Registry, found four previously accessioned cases which had been unrecognized. These mistaken diagnoses are worthy of note.

Apparently the first case to be accessioned in the Registry was that of Rodin which was entered in 1928 (Case 4). This was diagnosed as a hemangioma of the iris and was so reported by Rodin[19] in 1929 as the third such case in the literature. This case probably represents the first reported instance of intraocular nevoxanthoendothelioma. This case was also used by DeCoursey and Ash in their *Atlas of Ophthalmic Pathology*, published in 1939,[20] as an example of hemangioma of the iris. The second case was added in 1937 by Friedenwald, diagnosed as malignant melanoma of the iris (Case 5). Case 6 was accessioned in 1945, diagnosed as chronic iridocyclitis. This case is particularly interesting because of the comment of Dr. Frederick Verhoeff, "To me this is an extremely puzzling specimen. . . . If these are primary tumors of the iris they are unique." Case 11 was accessioned in 1949 with the pathologic diagnosis of an unusual form of neurofibroma. As noted above, Case 7 was originally diagnosed as malignant melanoma in 1938.

PATHOLOGY

The pathologic lesion of the skin nodule is usually quite characteristic and can be diagnosed by the trained observer in a large percentage of cases (Figure 1). This lesion consists of a small, circumscribed, nodule of tightly packed cells in the corium. The overlying epithelium is usually normal except for slight atrophy. The nodule consists of a mass of large mononuclear cells, many of which are polygoneal or spindle-shaped. The cytoplasm is usually clear, but some cells contain fat, causing a foamy appearance (Figure 2). The presence of Touton-type giant cells is characteristic, this cell being made up of lipid cytoplasm rimmed by nuclei. There is often an occasional eosinophile. There is usually a diffuse groundwork of connective tissue and at times there may be some new-formed blood vessels.

The eye lesion consists of a diffuse infiltration of the iris with a similar large histocyte (Figure 3). This infiltration is often so dense as to replace the normal iris structure. This infiltration often extends into the ciliary body, apparently involving it secondarily. The large histocytes may also be seen on the surface of the ciliary processes but are

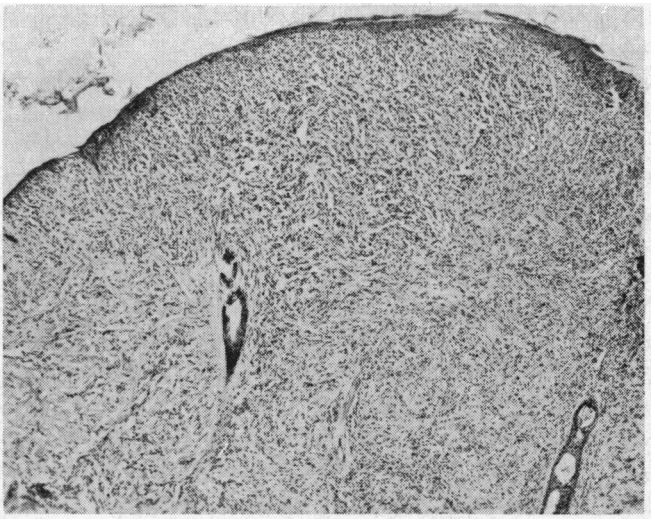

FIGURE 1. BIOPSY OF SKIN LESION (LOW POWER)

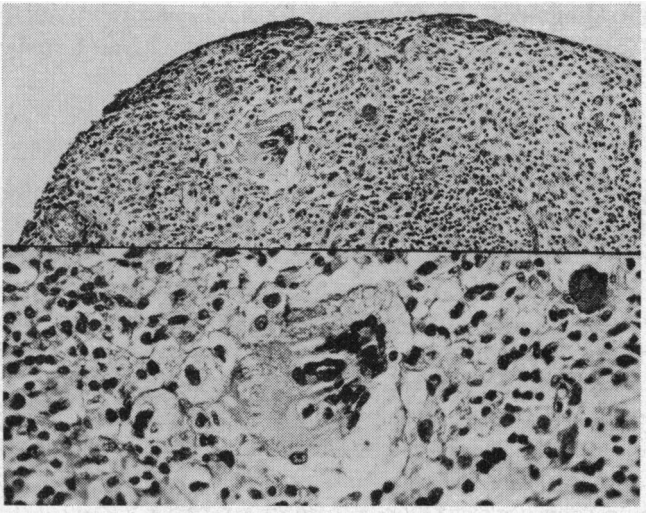

FIGURE 2. BIOPSY OF SKIN LESION—HISTIOCYTIC INFILTRATION AND TOUTON GIANT CELL

rarely seen in the vitreous itself. In no case has any posterior involvement been noted.

The essential cell of the iris lesion is a large monocyte with clear eosinophilic cytoplasm (Figure 4). When in tissue they may be

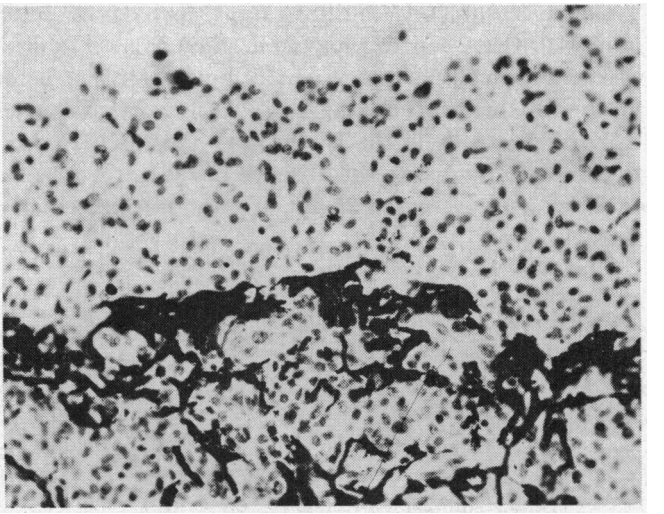

FIGURE 3. INFILTRATION OF IRIS STROMA WITH HISTIOCYTES

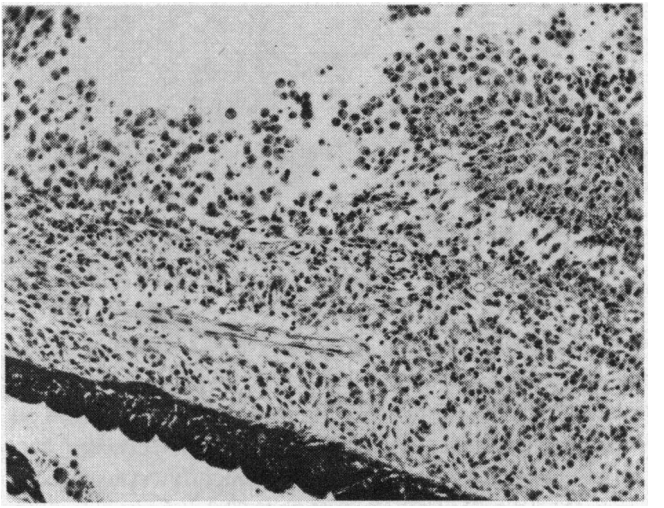

FIGURE 4. SURFACE OF IRIS SHOWING SHEDDING OF THE TYPICAL HISTIOCYTES

polygoneal, but when free on the surface of the iris or in the aqueous they appear to be a large, round cell without distortion. They tend to form a layer on the anterior surface of the iris which often appears to be shedding into the aqueous (Figure 4). These cells float into the

aqueous and may completely infiltrate the trabecular structures in a manner similar to that seen in phacolytic glaucoma (Figure 5). The Touton-type of giant cell may occasionally be present but is much less frequent in the iris than in the skin. Also it is noted that there is much less tendency for the cells in the iris to have the foamy, lipid type of cytoplasm. This may be due to the absence of fat in the iris. Occasionally eosinophiles may be noted in the iris lesion, particularly in the earlier stages.

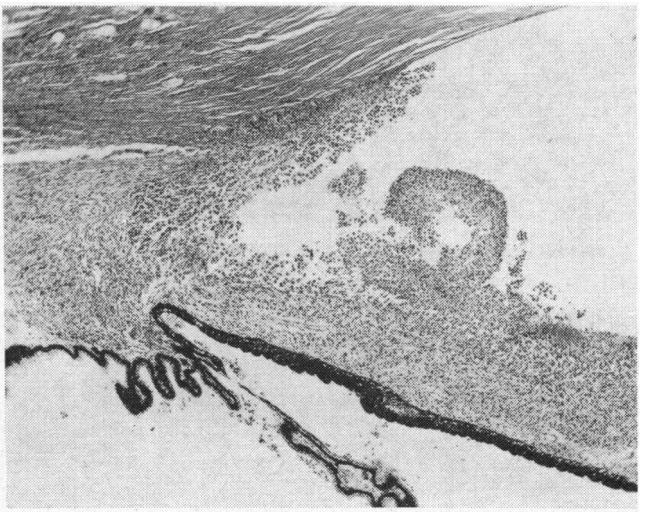

FIGURE 5. INFILTRATION OF ANGLE WITH HISTOCYTES WITH NODULE ON IRIS SURFACE

In the stroma many large, new-formed, thin-walled blood-vessels are often present and seem to be characteristic of the iris lesion (Figure 6). This accounts for the mistaken diagnosis of Rodin's case as hemangioma. This characteristic of the iris lesion is surely the cause of the anterior chamber hemorrhage that is so clinically typical of the disease. Old blood with secondary fibrosis at times may be present on the iris.

As noted, the glaucoma is caused by a cellular infiltration of the intratrabecular spaces. The anterior chamber hemorrhage may add to the trabecular block. Later peripheral anterior synechia may form. In the advanced cases of glaucoma, the iris may undergo complete atrophy with some slight secondary fibrosis. Even in these advanced

cases, an occasional histocyte may still be present. In these cases the typical findings of infantile glaucoma are present in the posterior segment (Cases 1 and 2).

The similarity of the lesion in the skin and in the iris is striking but probably to be expected. This similarity is seen not only in the cytology of the histocytic infiltration, but also in the granulomatous characteristics

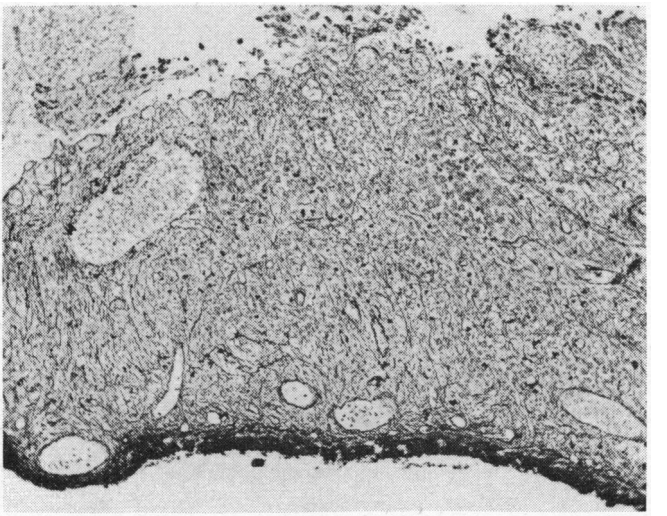

FIGURE 6. NEOVASCULARIZATION OF IRIS STROMA

with the presence of the Touton giant cell. The secondary change of the neovascularization is present in both, but secondary fibrosis is minimal in the iris, giving rise to a more diffuse lesion. On the iris surface and in the anterior chamber the histocytic proliferation is particularly different due to the absence of restraining tissue barriers.

One of the most obvious peculiarities of the disease is the apparent limitation of the lesions to the stroma of the skin and the iris, but any lesion in these sites is easily recognized clinically. However, this localization may suggest some common but undetermined characteristic of these sites that cause them to be susceptible to the lesion. It is possible that there may be some subclinical sites that are not recognized in the absence of death and autopsy. Helwig and Hackney[7] have reported lesions in the lung and testes in an autopsied case.

CLINICAL CHARACTERISTICS

This disease is characterized clinically by the presence in an infant of an iris lesion with a high incidence of spontaneous anterior chamber hemorrhage and severe secondary glaucoma (Figure 7). The skin lesion, if present, may be diagnostic both clinically and histologically (Figure 8).

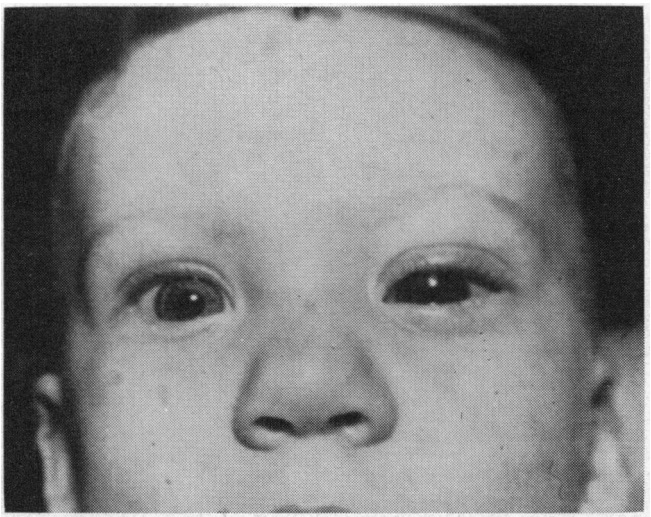

FIGURE 7. CASE 1 SHOWING HYPHEMA, MEGALOCORNEA, AND SKIN LESION ON FOREHEAD

One of the most characteristic features of the disease is the age incidence, as in all cases in this series except three, the onset of the disease was under the age of one year. It is even more striking that nine of the patients were six months or under at onset. Of the three older patients the onset occurred at one and a half years, three years, and 35 years. This one adult is the only exception in our series that this is a disease of infants and young children. Helwig[21] has stated that the skin disease does occur rarely in adults. It was to be expected that an iris lesion in an adult would eventually be recognized. Possibly because of early recognition this case lacks some of the typical clinical findings.

Helwig and Hackney[7] state that the sexes in their series were about evenly divided. This is true in the present series with 10 males and 10 females. All patients were white except Case 8.

In all patients a gross iris lesion was present clinically. In the majority of cases the iris is described as being diffusely thickened with a muddy discoloration of the stroma. This may cause a gross heterochromia of the iris. In three patients a localized iris mass was noted. The examination of the anterior chamber may be difficult because of the steamy cornea and the presence of hyphema.

FIGURE 8. SKIN LESION FROM FOREHEAD

Probably the most important clinical finding is the hyphema. The original iris lesion is often not appreciated and the presence of the blood in the anterior chamber is the first finding noted by the parents. Frank anterior chamber hemorrhage was noted in 17 patients, being absent in only two, one of these being the adult. In at least four patients the hyphema was recurrent.

Glaucoma is a common finding and the increased pressure may come on rather abruptly with a rapid enlargement of the cornea. Glaucoma is reported being present in 16 patients and absent in only two, one of these being the adult. One case had goniotomy and cyclodiathermy (Case 12), both of which were unsuccessful. Newell's first case[16] had a successful filtration operation for the increased tension and the globe was retained. Case 15 had a trephining about one year after the iridectomy. In two other patients (Cases 1 and 2) in whom the diagnosis was made clinically and by skin biopsy, the eye was retained for

as long as three years (Case 2), but was subsequently enucleated because of the marked enlargement of the globe with pain.

The skin lesion is usually quite characteristic clinically as well as histologically. Skin lesions are noted as being present in eight of these cases. In only three patients was the skin lesion absent when searched for, one being the adult. In the majority of the remaining cases, particularly the earlier ones, the lesions possibly were, if present, not noted or the interrelationship of skin and eye lesions ignored. In the eight patients with skin lesions, seven of them had diagnostic biopsies, the remaining patient (Case 3) being typical clinically. One case (Case 13) was diagnosed on the biopsy of the skin lesion without iris biopsy. In two patients (8 and 13) the iris lesion preceded the skin manifestations by an interval of some weeks.

The skin lesions consist of a small, circumscribed, elevated papule usually yellowish in color, but it may be pinkish in the early stage (Figure 8). They are usually multiple. They are more commonly found on the scalp and head, but may be present on the trunk and extremities. In Helwig and Hackney's[7] series one-fifth were present at birth. Skin lesions tend to undergo spontaneous regression over a long period and may leave a slightly umbilicated scar for a number of months which eventually completely disappears.

DISCUSSION

The term nevoxanthoendothelioma, as suggested by McDonaugh[4] in 1909, has become the usual designation for this disease, particularly in the dermatologic literature. It is cumbersome and is incorrect in the light of our present knowledge of the lesion, juvenile xanthogranuloma, as suggested by Helwig and Hackney,[7] is definitely to be preferred.

There is some disagreement in the literature as to the exact relationship of nevoxanthoendothelioma to the other lipoidal histiocytoses, or the reticuloendothelioses. This group includes eosinophilic granuloma of bone, Hand-Schuller-Christian disease, and Lederer-Siwe disease. These diseases were grouped together by Lichtenstein in 1953[22] under the diagnostic term "Histiocytosis X." Each of these diseases in its pure form is a rather distinct entity both clinically and pathologically. However, it is well accepted that transitional forms are often encountered.[22] A common feature to both the reticuloendotheliosis group and nevoxanthoendothelioma is an apparently normal lipid metabolism. In each condition the etiology is unknown.

Nevoxanthoendothelioma has been put into this group by some writers[9, 15, 16, 23-25] because of a number of histopathological similarities between the lesion of nevoxanthoendothelioma and that of the reticuloendotheloses. There is excellent evidence to support the belief that nevoxanthoendothelioma is an entirely different entity and is probably a unique disease, this idea being supported particularly by Helwig[26] and Nomland.[11]

We are inclined to support this belief because of a number of reasons. (1) Although similar, there is probably some distinct and diagnostic histologic differences between the skin lesion of nevoxanthoendothelioma and the reticuloendotheloses.[27] (2) Iris lesions such as seen in nevoxanthoendothelioma have not been reported in the reticuloendotheloses. (3) The lack of transition forms between the histiocytosis X group and nevoxanthoendothelioma. No case of nevoxanthoendothelioma has been reported which has merged into the other group. (4) The apparent lack of visceral lesions other than that of the iris is characteristic of nevoxanthoendothelioma while the lesions of the reticuloendotheloses tend to be multiple. As noted, it is possible that visceral lesions of nevoxanthoendothelioma are not determined in the absence of autopsy. (5) The benign course of nevoxanthoendothelioma with spontaneous regression is quite different than the severe course, often fatal, of the reticuloendotheloses.

Iris lesions seem to be the only common visceral complication in nevoxanthoendothelioma. Although much rarer than the skin manifestation, it is probably the most important aspect of this disease. The skin lesion runs a benign course, undergoing spontaneous regression, while the presence of the iris lesion is often fatal to the eye. The reason for localization of this peculiar secondary lesion in the iris is unknown.

The tendency for spontaneous regression which occurs so regularly in the skin probably also occurs in the iris. It is interesting to speculate that there may be cases of subclinical nevoxanthoendothelioma which could be unrecognized. This would be true if the skin lesions were not noted and the iris lesion so mild that the complications of glaucoma or anterior chamber hemorrhage did not occur. It is also interesting to note that no bilateral case has yet been recorded.

It is obvious that the intraocular involvement of nevoxanthoendothelioma is much more common than was supposed a short while ago. Prior to 1955 only one case of intraocular involvement had been recognized and reported. Since that time over 20 cases have been reported and probably a number of others have been recognized clinically but

not reported. The 20 cases in this series show an amazing similarity in all features both clinical and pathologic.

The clinical picture of this disease, consisting of a diffuse iris lesion and spontaneous anterior chamber hemorrhage with severe secondary glaucoma in an infant, should be familiar to every ophthalmologist so that a prompt and exact diagnosis can be made. Spontaneous anterior chamber hemorrhage in an infant from any other cause is a rarity. With this finding alone the diagnosis of nevoxanthoendothelioma should be suspected. In all such cases the careful search of the skin should be instituted so that any skin lesion present can be biopsied to substantiate the diagnosis. This would prevent the needless enucleation of these eyes for suspected intraocular tumor. In those cases of the eye syndrome without skin lesions, iris biopsy is probably indicated.

The prognosis of this disease is not as poor as suggested by this series, as 13 eyes were enucleated because of the mistaken diagnosis of intraocular tumor. With improved recognition this should not occur. It is significant that in four of the more recent cases the lesion was diagnosed by iris biopsy and the globe retained. At present the follow-up in these biopsy cases is insufficient to determine the final result. However, even with a correct diagnosis, the eye will be lost if the glaucoma is not controlled (Cases 2 and 3). Any treatment to be successful must suppress the histiocytic proliferation so that the complications of secondary glaucoma and anterior chamber hemorrhage will not develop. If this can be accomplished even temporarily, spontaneous regression of the iris lesion will probably occur later.

Only recently has any promising therapy been suggested. Maumenee and Longfellow[18] have reported two cases treated with X-ray therapy with interesting results. They believe, in view of the type of lesion present in the iris and the success of X-ray therapy in similar lesions elsewhere, that this treatment is logical. Hedges[17] has added another case with complete disappearance of the lesion although followed only three months. Hogan[28] has also successfully treated a case. Maumenee and Longfellow[18] suggest a preliminary trial of topical and systemic steroid therapy. In our first two cases topical steroids were used without benefit. Maumenee and Longfellow[18] believe that X-ray therapy should be used only if steroids fail to control the glaucoma, which may also be treated by Diamox and miotics. They emphasize that a single dose of not more than 200 r be given so as to prevent the formation of cataract, which is the most feared complication of this therapy. However, cataract is to be preferred to severe secondary glaucoma with eventual loss of the globe. Early recognition of future cases with prompt radiation will allow this therapy to be properly evaluated.

SUMMARY

1. Twenty cases of intraocular nevoxanthoendothelioma, or juvenile xanthogranuloma, are reported, suggesting that the lesion is more common than is supposed.

2. Juvenile xanthogranuloma is a more correct and preferable designation for this disease.

3. The pathology of the lesion in both iris and skin is characteristic of this condition and explains the clinical findings.

4. The clinical picture of the iris tumor, spontaneous anterior chamber hemorrhage, and secondary glaucoma in an infant should suggest the diagnosis of this disease and indicate a search for skin nodules.

5. It is believed that this disease is unique and is unrelated to the reticuloendotheloses.

6. Although the management in the past has usually been unsatisfactory, X-ray therapy is promising. It is suggested that it be further evaluated in this condition.

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DISCUSSION

DR. W. E. FRY. I am pleased to be able to discuss this paper of Dr. Sanders. His survey makes a valuable addition to our knowledge concerning intraocular nevoxanthoendothelioma. It is also an interesting comment on the fact that when an unusual condition is first recognized, additional cases rapidly are documented in the literature.

I had the opportunity of examining the eye that has proved to be the first case with an intraocular complication. I should like to refer to that case. This was reported by Blank, English, and Beerman in the pediatric literature in 1949. The eye sections of this case were demonstrated at the meeting of the Ophthalmic Pathology Club in 1948. A report of the case was made by Dr. Nachaud at the meeting of the College of Physicians in Philadelphia on December 16, 1948. It was discussed by Drs. Fewell, John Scott, and Herman Beerman and was recorded in the *American Journal of Ophthalmology*, December, 1949, in the Proceedings of Societies.

The patient was a four-month-old boy admitted to the service of Dr. Fewell at the Children's Hospital and was cared for by one of his associates.

The history is that of an inflamed painful eye of five days' duration. On admission, the cornea was hazy, the iris seen indistinctly, but on it a pigmented mass could be seen. No fundus view was obtained, the tension was elevated, transillumination was poor. After observation and treatment for a period of a week, the eye was enucleated. There were no skin lesions at this time.

The pathologic sections of the eye revealed unusual changes in the iris that were not typical of either neuroepithelioma or sarcoma.

It was not until four months later that skin lesions were noted on the head and trunk. These were biopsied and a diagnosis of nevoxanthoendothelioma was made and comparison with the iris lesion indicated the same involvement so that a diagnosis of intraocular involvement could be confirmed.

Enucleation of the eye was justified by the finding of a secondary glaucoma, initiated in part by the infiltrating lesion and a total dislocation of the lens into the vitreous cavity through a tear in a detached retina to become lodged on the choroid in the subretinal space.

The histologic features that should be emphasized are the dense infiltration of cells similar to large histocytes, the presence of Touton giant cells, and the tendency of the infiltration to overflow the borders of the iris tissue.

The important clinical features are the presence of a diffuse iris lesion with a hyphema in a child with or without secondary glaucoma. When these are present, the possibility of a nevoxanthoendothelioma must be considered.

The possibility of resolution either spontaneously or following radiation needs to be noted so that enucleation of some of these eyes can be avoided.

DR. A. EDWARD MAUMENEE. I should like to congratulate Dr. Sanders on his very excellent summary and presentation of the 20 most quoted unpublished cases in the literature. We have been expecting this paper for a number of years, and I am delighted to see he has finally gotten these cases together for presentation.

One of the most interesting findings in this group of cases was a patient who was 35 years of age. This is certainly unique in juvenile xanthogranuloma. It suggests that we should look for this lesion in some of the rare and unusual types of uveitis in adults.

The problem that I should like to speak of for the moment is the treatment of these cases. The eyes with this disease are usually lost because of glaucoma; therefore it is most important to give the child a general anesthesia or sedate him with a lytic cocktail to check the tension. If it is not elevated, the child should be followed. If the tension is elevated, Diamox should be instituted and systemic cortisone tried. In the reticuloendotheliomas, such as Christian-Schueller disease, there have been reports of remissions when systemic cortisone has been given. If the eye does not seem to respond to this conservative type of therapy, then X-ray therapy directed to the anterior segment of the eye is justified. I should like to give a word of caution, in that Dr. Merriam has reported that as little as 500 in one dose will produce a cataract in the eye of a child. Therefore, the therapy should be 100 roentgen units every week or two weeks until approximately 400 roentgen units had been given.

In 1954 we treated a juvenile xanthogranuloma of the iris in a three-year-old child. A diagnosis of the lesion had been confirmed by biopsy. Almost five years later, in 1958, the iris was slightly atrophic and darker than the other eye, there was a very slight lenticular opacity, but the visual acuity was 20/40.

DR. MICHAEL J. HOGAN. I have had the opportunity to see two cases of this condition. One has been reported by Dr. Thomas Hedges. His case was seen in consultation and the diagnosis was made on the basis of the clinical features. There were no skin lesions at this time. Based on previous discussions with Dr. Sanders, we took it for granted that the diagnosis in this case was nevoxanthoendothelioma, and the eye was treated with a total dose of 400 roentgen units of X-ray. The lesion responded rapidly, and Dr. Hedges has indicated after a two-year follow-up that it appears to be cured. The interesting thing in this case is that approximately six months after the diagnosis of the eye disease was made, there appeared a typical skin lesion and a biopsy showed a xanthogranuloma.

The second case was almost identical to the first. There were no skin lesions present at the time the eye lesions were observed. This case was also treated with 400 r tissue dose over a period of four weeks, as indicated by Dr. Maumenee, with an identical result. The child also developed skin lesions, three months after the completion of therapy. The child has been followed for a year now, without recurrence of the eye disease.

I think it is too bad the term nevoxanthoendothelioma is used for this disease, and we hoped that Dr. Sanders would use the preferred term juvenile xanthogranuloma. The lesion is not a nevus and it is not an endothelioma, and I do not see why we have to perpetuate the usage of a poor term.

DR. SANDERS. I wish to thank the staff of the Armed Forces Institute of Pathology, Dr. Lorenz Zimmerman, and Miss Eleanor Paul for assistance in collecting the clinical data and for the photomicrographs. I also wish to thank the discussers. I think it is rather unusual that, with one exception, everyone who has written on this condition is here today.

Dr. Maumenee, if I had written this when I threatened to several years ago, I should only have had ten cases. I think it worthwhile to have waited to get the other ten.

In both our cases we used topical stereoids with no effect. Because the patients were infants we felt large doses of systemic steroids were contraindicated. I think Dr. Maumenee may be right that glaucoma is the indication for X-ray therapy. However, the tension goes up very rapidly; so if one is not going to use X-ray immediately, one must watch for rising tension. I think Dr. Maumenee's and Dr. Hogan's discussions show X-ray therapy is the treatment of choice, a most valuable one, and should be used immediately.

I agree with Dr. Hogan that it is preferable to use the term juvenile xanthogranuloma, and this change in the original title has been made.